Photo essay

Oguchi's Disease With Mizuo-Nakamura Phenomenon

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Abstract

Oguchi's disease is an unusual form of congenital stationary night blindness with autosomal recessive inheritance. Mizuo-Nakamura phenomenon is classical of Oguchi's disease. Full-field electroretinogram shows absent rod response and essentially normal cone-mediated response. The mixed rod–cone response has a negative configuration with relatively well-preserved oscillatory potentials. It recovers to a near normal level after a long period of dark adaptation. We herein report a case of Oguchi's disease with Mizuo-Nakamura phenomenon.

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Keywords: *Oguchi's; Mizuo-Nakamura Phenomenon; Electroretinogram*

Oguchi's disease is an unusual form of congenital stationary night blindness with autosomal recessive inheritance. It is caused by mutation of arrestin or rhodopsin kinase.[1] We herein report a case of Oguchi's disease with Mizuo-Nakamura phenomenon.

A 22-year-old male came with complaints of night blindness since childhood. His visual acuity was 20/20 in both eyes. Color vision was normal. Anterior segment examination was unremarkable. Fundus examination of both eyes revealed diffuse golden, tapetal-like sheen with mild retinal vascular attenuation (Figures 1a and 1b) which was extinguished after 45 minutes of dark adaptation with normal visible retinal vasculature (Figures 1c and 1d). This characteristic phenomenon observed on fundus examination from dark-adapted state to light adaptation is known as Mizuo-Nakamura phenomenon. There was no evidence of bony spicule pigmentation. Full field electroretinogram showed non-recordable single flash rod response with characteristic negative waveform morphology in combined rod-cone response and normal cone response (Figure 2). A diagnosis of Oguchi's disease was made. Patient was advised for screening of family members and genetic counselling but patient was lost to follow up.

Mizuo-Nakamura phenomenon is classical of Oguchi's disease.[2] This can also be seen in retinitis pigmentosa,[3] X-linked retinoschisis[4] and cone-rod dystrophy.[5] It has been reported that it occurs due to elevated extracellular potassium levels generated in the retina in response to an excessive stimulation of rod photoreceptors. Thus, a patient of Oguchi's disease presents with characteristic clinical

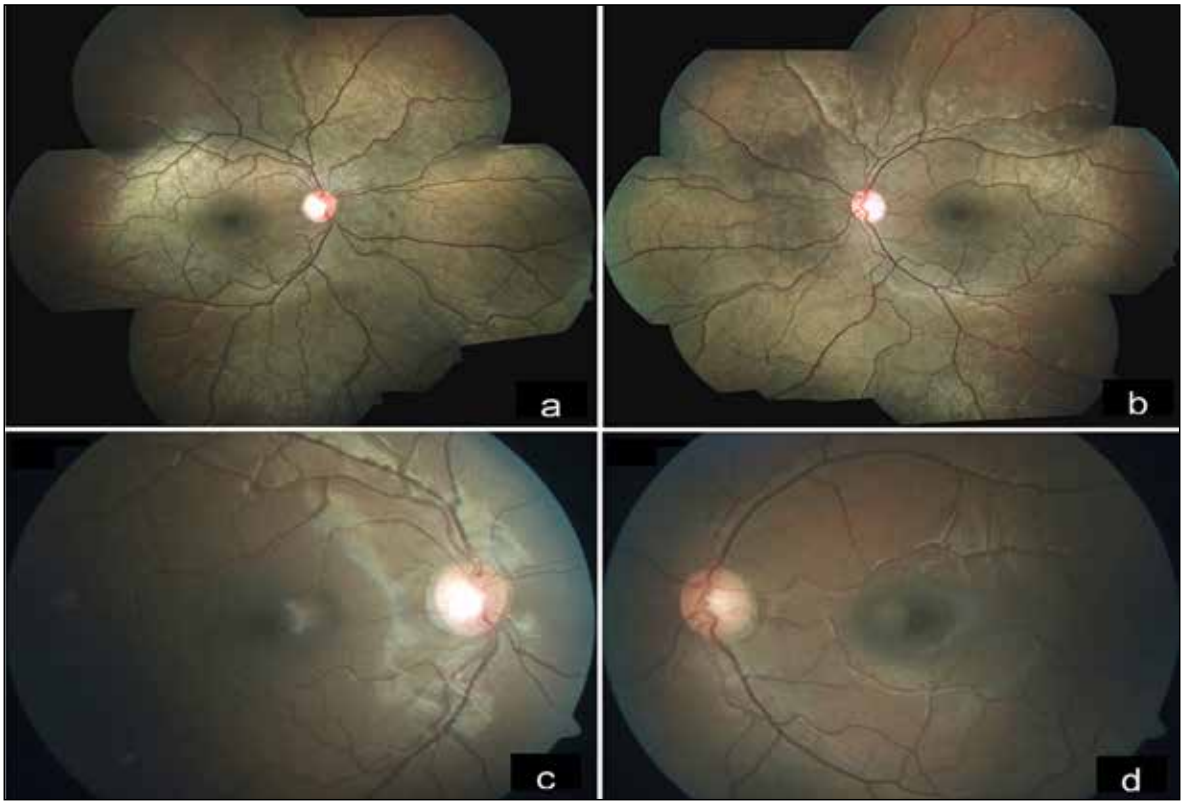

Figure 1: *Montage color fundus photograph of right (a) and left (b) eye reveal a golden tapetal-like sheen with mild retinal vascular attenuation that is extinguished after 45 minutes of dark adaptation with normal visible retinal vasculature suggestive of the Mizuo-Nakamura phenomenon as seen in the color fundus photograph of right (c) and left (d) eye respectively.*

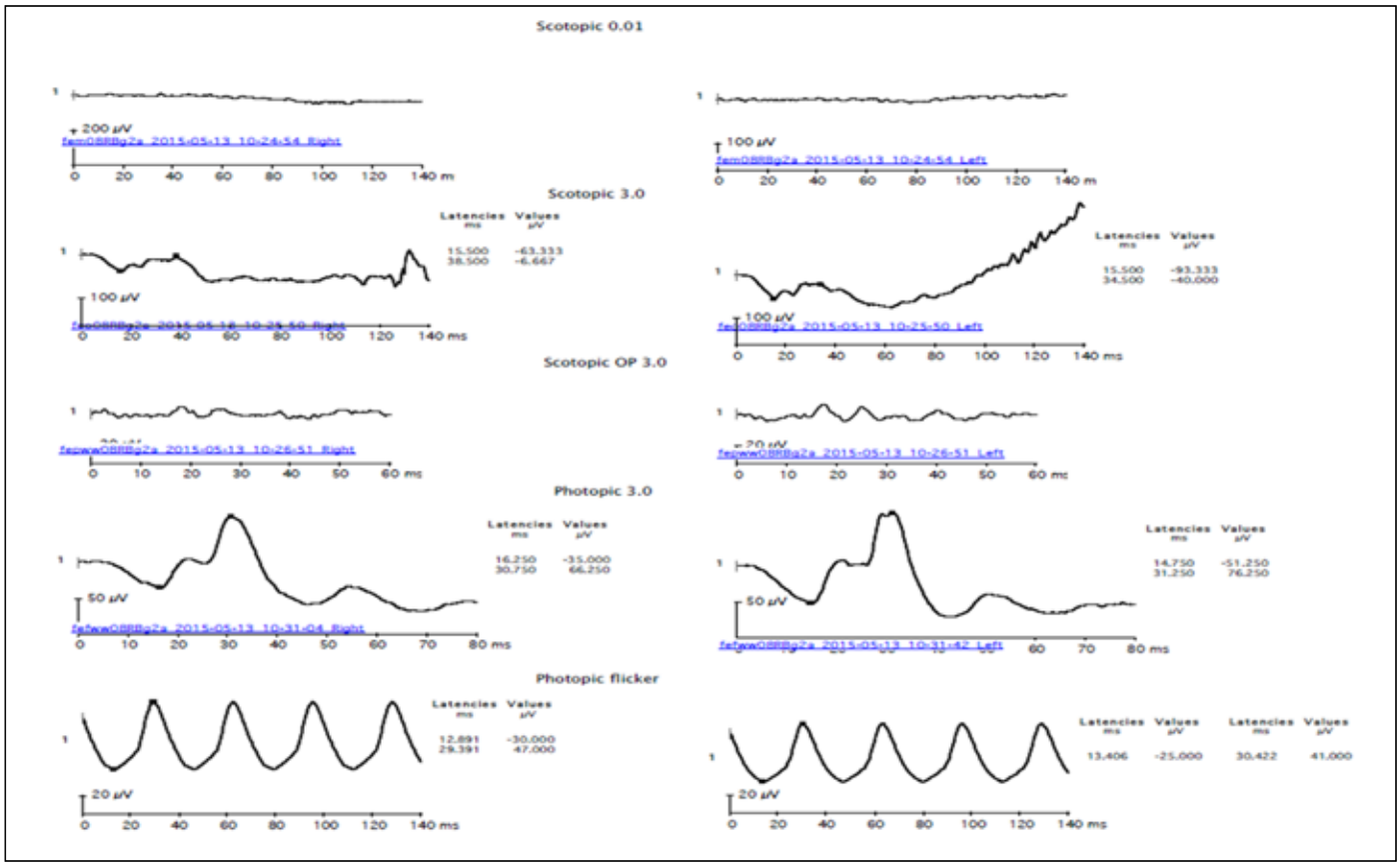

Figure 2: *Full field electroretinogram of both eyes showing non-recordable single flash rod response, negative waveform morphology in combined rod-cone response and normal photopic response.*

features. Electroretinogram corroborates the clinical findings. Full-field electroretinogram shows absent rod response and essentially normal cone-mediated response. The mixed rod–cone response has a negative configuration with relatively well-preserved oscillatory potentials. It recovers to a near normal level after a long period of dark adaptation.

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